Herlyn Werner Wunderlich syndrome (HWWS): an unusual presentation of acute abdominal pain

Giulia van der Byl · Vincenza di Giacomo ·
Vittorio Miele

Received: 26 March 2014 / Accepted: 28 March 2014 / Published online: 1 May 2014
© Società Italiana di Ultrasonologia in Medicina e Biologia (SIUMB) 2014

Herlyn Werner Wunderlich syndrome (HWW) is a very rare congenital malformation of the urogenital tract secondary to Müllerian developmental abnormalities MDA.

HWW syndrome results from agenesis or hypoplasia, defective vertical or lateral fusion, or resorption failure of paramesonephric ducts.

Anatomically it is characterised by a triad of didelphys uterus, obstructed hemivagina and ipsilateral renal agenesis (Fig. 1). This syndrome was described the first time on 1922 [1].

Didelphys uterus is generated by the absence of fusion of the Müllerian cavity, lacking of communication between the two uterine cavities, each hemiuterus show regular zonal anatomy, with its vagina, cervix, Fallopian tube and adnexa [2, 3].

Being obstructed the vaginal ostium, after menarche, at every menstrual cycle; blood material accumulates gradually into the vagina (hematocolpos), in the uterus (hematometra) and along the course of the ipsilateral tube (hematosalpinx). Some of the blood is absorbed between the periods, and therefore, patients with HWW syndrome can go unrecognised for several years after the onset of menstruation.

One other reason for delayed diagnosis is the presence of regular menses.

This anomaly is generally observed in post-menarche adolescents and young women with irregular menses, dysmenorrhoea, abdominal pain and pelvic mass. The diagnosis may be difficult due to the infrequency of this syndrome, and a high index of suspicion is required.

Reproductive performance of these patients is affected by the delayed diagnosis: retrograde blood flow destroys tubal function and leads to endometriosis [4]. Another important factor affecting fertility is the occurrence of infectious complications.

Radiologic imaging is fundamental tools in the diagnosis of HWWS. Ultrasound is the first step in the evaluation of undifferentiated pelvic pain; even though US is necessary to pounce the suspect of Müllerian anomalies, ultrasound is not able to clearly identify the anatomical abnormality in this patient. MRI further characterises structural abnormalities with high accuracy, and results necessary to plan for surgical management.

We report a case of a 10-year-old female who presented to our Emergency Department with severe abdominal pain that occurred, less intense, since the patient achieved menarche, 6 months before; the patient had also irregular cycle with severe dysmenorrhoea, and she reported that the pain begun immediately prior to menstruation and worsened the second and third day of the menstrual period. Abdominal examination revealed a palpable mass.

A transabdominal ultrasound examination was performed using a convex (3.5–5.0 MHz) and a linear probe (5.0–12.0 MHz).

US findings showed two separate uterine cavities and cervixes, suggestive of uterus didelphys (Fig. 2). The right uterine cavity and cervix were normal (Fig. 3). The left uterine cavity and cervical canal were dilated and filled

G. van der Byl (✉)
IRCCS Foundation, San Matteo Medical Center, Institute of
Radiology, University of Pavia, Viale Golgi, Pavia, PV, Italy
e-mail: giulivan@libero.it; giulia.vanderbyl@gmail.com

V. di Giacomo · V. Miele
Unità Operativa Diagnostica per Immagini nel DEA e per le
Urgenze, Azienda Ospedaliera S. Camillo-Forlanini,
Circonvallazione Gianicolense 87, Rome, Italy
e-mail: enzadigiaco1983@hotmail.it

V. Miele
e-mail: vmiele@scamilloforlanini.rm.it

Fig. 1 The drawing illustrates anatomical variations (renal agenesis, didelphys uterus and obstructed hemivagina) and consequences of the HWW: hematometra, hematosalpinx and hematocolpos on the same side of the renal agenesis; on the opposite side, the zonal anatomy is preserved

Fig. 2 US shows dilated left hemivagina, Fallopian tube and hemiuterus. On the right side is recognisable the right normal hemiuterus (*white arrow*). Axial turbo spin-echo T1-weighted fat-saturated MR image US shows analogous findings

Fig. 3 US sagittal scan shows right uterine cavity with normal cervix and vagina (*white arrow*); same findings are showed in the sagittal T2W MRI scan

with fluid that was hypoechoic with few hyperechoic areas (Fig. 4). In addition, a tubular structure was noted in left adnexa location extending laterally from left uterine corn with fluid within, suggestive for left hematosalpinx (Fig. 5). Right and left ovaries were normal (Fig. 6). In addition, renal agenesis of the left kidney (Fig. 7) with compensatory hypertrophy of the right kidney was identified.

MRI was performed to confirm the hypothesis of Müllerian duct anomalies and to precise anatomical delineation of the uterus, tubes, cervix and vagina. Status of these reproductive organs and extent of endometriosis, pelvic inflammation and adhesions can be well evaluated with MRI.

MRI showed two separate uterine cavities, cervixes and vaginas. Normal zonal anatomy was revealed on the right side.

Left uterine cavity, cervical canal and salpinx were dilated and filled with fluid hyperintense on both T1w and T2w images with few T2 hypointense areas suggesting blood products.

Fig. 4 US shows centrally the vagina (V) and laterally on the left the Fallopian tube (T) and the hemiuterus (U) filled by inhomogeneous hypoechoic fluid. Axial turbo spin-echo T1-weighted fat-saturated MR image confirms the findings: vagina, left tube and hemiuterus are distended by inhomogeneous hyperintense fluid, signal due to products of haemoglobin degradation

Fig. 5 US shows in the left hemipelvis, over the bladder a tubular structure dilated and filled with inhomogeneous hypoechoic fluid; behind it is recognisable the left hemivagina filled by inhomogeneous material (arrow). Sagittal T2-weighted MR image confirms the

findings: vagina and the left tube are distended by inhomogeneous hypointense fluid; coronal turbo spin-echo T1-weighted fat-saturated MR images show the haematosalpinx

Fig. 6 US shows dilated left hemivagina and Fallopian tube. The left normal adnexa (white arrow) is located in front. Analogous findings are showed in axial T2W MRI scan

Fig. 7 US coronal image through the left flank reveals absent kidney

The presence of hematocolpos, hematometra and hematosalpinx supported the hypothesis of being an obstructing left vaginal septum.

At examination under general anaesthesia, an obstructed left hemivagina was identified with a longitudinal vaginal septum. A vaginal septectomy was performed, and a large amount of haematic fluid was drained.

Conflict of interest Giulia van der Byl, Vincenza di Giacomo, Vittorio Miele declare that they have no conflict of interest.

Informed consent All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000 (5). All patients provided written informed consent to enrolment in the study and to the inclusion in this article of information that could potentially lead to their identification.

Human and animal studies The study was conducted in accordance with all institutional and national guidelines for the care and use of laboratory animals.

References

1. Purslow CE (1922) A case of unilateral haematocolpos, haematometra, and haematosalpinx. *J Obstet Gynaecol Br Emp* 29:643
2. Troiano RN, McCarthy SM (2004) Mullerian duct anomalies: imaging and clinical issues. *Radiology* 233(1):19–34 (Epub 2004 Aug 18)
3. Junqueira BL, Allen LM, Spitzer RF, Lucco KL, Babyn PS, Doria AS (2009) Müllerian duct anomalies and mimics in children and adolescents: correlative intraoperative assessment with clinical imaging1. *Radiographics* 29(4):1085–1103. doi:[10.1148/rg.294085737](https://doi.org/10.1148/rg.294085737)
4. Güdücü N, Gönenç G, Işçi H, Yiğiter AB, Dündar I (2012) Herlyn-Werner-Wunderlich syndrome—timely diagnosis is important to preserve fertility. *J Pediatr Adolesc Gynecol* 25(5):e111–e112. doi:[10.1016/j.jpog.2012.05.013](https://doi.org/10.1016/j.jpog.2012.05.013) (Epub 2012 Jul 27)